

Citrullinemia Care Plan (last updated 2/21/09)

<p>Clinical Considerations</p> <ul style="list-style-type: none"> • Stabilizing neonate (meds & hemodialysis) 	<p>Initial labs (diagnostic & baseline)</p> <ul style="list-style-type: none"> • Quant plasma amino acids • CMP and ammonia • Consider blood gases, UOA, urine orotic acid, PT/PTT • Consider Argininosuccinate synthase (ASS) enzyme assay on fibroblasts for CIT • Consider sequencing for CIT 												
<p>Diet considerations/ treatment</p> <ul style="list-style-type: none"> • Low-protein diet • Formula (essential AA) • Medication for hyperammonemia <ul style="list-style-type: none"> ○ Carnitine reported in 1 paper • Avoidance of fasting • Liver transplant 	<p>Monitoring Quant plasma AA</p> <table border="1" style="width: 100%; border-collapse: collapse; text-align: center;"> <thead> <tr> <th style="padding: 2px;">Age</th> <th style="padding: 2px;">Frequency</th> </tr> </thead> <tbody> <tr> <td style="padding: 2px;">0-6 months</td> <td style="padding: 2px;">Every 2 weeks</td> </tr> <tr> <td style="padding: 2px;">6-12 months</td> <td style="padding: 2px;">Monthly</td> </tr> <tr> <td style="padding: 2px;">1-6 years</td> <td style="padding: 2px;">Every 3 months</td> </tr> <tr> <td style="padding: 2px;">6-18 years</td> <td style="padding: 2px;">Every 6 months</td> </tr> <tr> <td style="padding: 2px;">>18 years</td> <td style="padding: 2px;">Yearly</td> </tr> </tbody> </table>	Age	Frequency	0-6 months	Every 2 weeks	6-12 months	Monthly	1-6 years	Every 3 months	6-18 years	Every 6 months	>18 years	Yearly
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<p>Emergency management</p> <ul style="list-style-type: none"> • Immediate 10% (or >) dextrose with salts + intralipids (may need insulin) • If ammonia high, IV meds or dialysis • Track ammonia levels, IV meds (dose), dialysis, central line, intubation, ICU care, days in hospital, coma score • Transfer to peds center if necessary 	<p>Labs to obtain during illness</p> <ul style="list-style-type: none"> • Comp metabolic panel • Ammonia • SAA 												
<p>Other evaluations</p> <ul style="list-style-type: none"> • CT /MRI scan at time of Dx if clinically warranted • MRI at 9 y or sooner if clinically warranted • Liver function • Bone health <ul style="list-style-type: none"> ○ DEXA-spine @9y for non-weight bearing & 18y for all others • Yearly developmental questionnaires (to be completed by parents) • Developmental eval @ 3 & 6y • Neuropsych @ 9y • Psychiatric screening @ 18 y • Referral to Liver and/or Renal as needed • Referral to Neurology as needed • Metabolic dietitian eval (at least yearly) 	<p>Yearly labs</p> <ul style="list-style-type: none"> • Comp metabolic panel with LFTs • PT/PTT • Consider ammonia • Prealbumin / albumin • Plasma Ferritin, transferrin, or iron studies • Consider CBC, hemoglobin, and hematocrit • Consider folate and vitamin B12 • Consider other nutritional testing (Zinc, selenium, vit D, essential fatty acids, and lipid profile). 												

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<p>Performance Measures</p> <ol style="list-style-type: none"> 1. Age of diagnosis (both positive NBS and confirmatory SAA) 2. Presence of illness at time of diagnosis including lethargy, somnolence, refusal to feed, vomiting, tachypnea, increased neuromuscular tone, spasticity, and ankle clonus. 3. Days until ammonia is within treatment range 4. Therapy during initial care <ol style="list-style-type: none"> a. Track IV meds (dose), dialysis, use of central line, intubation, ICU care, days in hospital, and coma score 5. Initial lab studies <ol style="list-style-type: none"> a. NBS results b. Quant plasma amino acids c. CMP d. Ammonia e. Blood gases f. UOA g. Urine orotic acid h. PT/PTT i. Molecular studies j. Enzymatic studies 6. Monitoring lab studies <ol style="list-style-type: none"> a. Quantitative serum amino acids b. Ammonia levels c. Nutrition labs 7. Frequency of clinic contacts and visits (track compliance with visits) 8. Total decompensations and hospitalizations. <ol style="list-style-type: none"> a. Track ICU admissions b. # of days of hospitalization c. # of ER visits d. Track ammonia level, IV meds (dose), dialysis, use of central line, intubation, and coma score 9. Growth parameters (ht, wt, OFC, BMI) 10. DEXA results and number of fractures 11. Diet <ol style="list-style-type: none"> a. Frequency of Metabolic dietitian visits (phone and clinic visits) b. Frequency of dietary analysis (3 day diet records) c. Natural protein intake (tolerance) d. Medical foods (formula)- (Y/N) e. Modified low-protein products (Y/N) f. Mode (oral, G-tube, bolus/drip, meds only/meds and diet) 	<ol style="list-style-type: none"> 12. Medications (include dosage) 13. Transplant (Y/N) 14. Neuropsychological evaluation 15. Psychiatric screening 16. Developmental services (PT, OT, & speech) 17. School performance <ol style="list-style-type: none"> a. Grade appropriate (Y/N) b. Special services (Y/N) c. IEP (Y/N) 18. Genetic Counseling (Y/N) <p>Outcome measures</p> <ol style="list-style-type: none"> 1. Mortality 2. Development <ol style="list-style-type: none"> a. IQ b. Level of functioning 3. History and/or presence of osteopenia 4. History and/or presence of abnormal MRI findings 5. History and/or presence of liver dysfunction 6. History and/or presence of psychiatric illness 7. Outcome of liver transplantation 8. Growth <ol style="list-style-type: none"> a. Final adult parameters
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